**Libyan Med J.** 2026;18(5):207-210
https://doi.org/10.69667/lmj.26501
**Libyan Medical Journal**
https://lmj.ly/index.php/ojs/index *eISSN: 2079-1224*

*Review article*

# Pseudomass-like CNS Lesions in Neuro-Behçet's Syndrome: A Narrative Review of a Rare Radiological Entity and Its Diagnostic Challenges

**Seraj Ajaj[1], Faisal Taleb[1]*, Abdussalam Abograra[2], Badar Allagi[3]**

[1]*Department of Neurosurgery, Tripoli University Hospital, Tripoli, Libya*
[2]*Department of Neuroradiology, Hamad Medical Corporation, Doha, Qatar*
[3]*Department of Neuroradiology, Tripoli University Hospital, Tripoli, Libya*
***Corresponding Email***. talebfaisal@gmail.com

***Keywords.***
*Neuro-Behçet's Syndrome, pseudomass, MRI, cerebellum, inflammatory vasculitis, Tumefactive lesion*

**ABSTRACT**

Neuro-Behçet's syndrome (NBS) is an uncommon but potentially disabling manifestation of Behçet's disease. While typical parenchymal NBS predominantly affects the brainstem, basal ganglia, and diencephalon, a rare subset presents as tumefactive or pseudomass-like lesions that mimic neoplasm, infection, and demyelinating disorders. These lesions create substantial diagnostic uncertainty and may lead to unnecessary neurosurgical intervention if misinterpreted. This narrative review synthesizes published evidence on pseudomass-like NBS, focusing on clinical context, neuroimaging patterns, pathological correlates, differential diagnosis, and treatment response to immunosuppressive therapy. A representative clinical vignette is included to illustrate typical radiological evolution and regression after therapy. Improved awareness of this rare radiological entity may facilitate timely diagnosis, avoid invasive procedures, and improve outcomes.

## Introduction

Behçet's disease is a chronic, relapsing, multisystem inflammatory vasculitis characterized by recurrent oral and genital ulcers, ocular involvement, and systemic manifestations. Neurological involvement, referred to as neuro-Behçet's disease (NBD), occurs in approximately 5–10% of patients, although prevalence varies by geographic region and cohort characteristics [1–3].

NBD is classically divided into parenchymal and non-parenchymal forms. The parenchymal form predominantly affects the brainstem, basal ganglia, thalamus, and deep white matter, whereas the non-parenchymal form most commonly manifests as cerebral venous sinus thrombosis [2–4]. Rarely, NBD may present as a mass-like or tumefactive lesion, also termed pseudotumoral or pseudomass NBD. These lesions often resemble neoplasms, abscesses, or granulomatous diseases on neuroimaging, posing a substantial diagnostic challenge and occasionally leading to unnecessary neurosurgical intervention [5–7]. Given the rarity of this presentation, evidence is largely limited to case reports and small series. A narrative synthesis of available data may assist clinicians in recognizing and managing this unusual manifestation. This narrative review synthesizes published reports on pseudomass-like or tumefactive presentations of neuro-Behçet's syndrome. The available evidence primarily comprises case reports, small case series, and narrative or pooled reviews. Due to the rarity of this entity and variability in reporting, findings are presented descriptively, with particular focus on radiological patterns, diagnostic challenges, and therapeutic responses, rather than through formal systematic methods or quantitative meta-analysis.

### *Epidemiology and Clinical Features*

In a pooled analysis of 43 patients with pseudotumoral NBD, the median age was approximately 36 years (range 12–59 years), with a male predominance of around 65–70% [6]. Unlike classical NBD, in which neurological manifestations typically occur several years after BD onset, pseudotumoral lesions may represent the inaugural manifestation of the disease. In the same review, more than half of patients had no prior diagnosis of BD at the time of CNS presentation [6]. Clinical manifestations are heterogeneous and include headache, focal neurological deficits, cerebellar signs, behavioral changes, seizures, and symptoms of raised intracranial pressure [5–8]. Notably, typical mucocutaneous or ocular features of BD may be absent or subtle, further complicating diagnosis [5, 7].

### *Neuroimaging Features and Lesion Distribution*

Neuroimaging findings in pseudotumoral NBD are variable but show recurring patterns. Lesions most frequently involve the capsulo-thalamic region, brainstem, and basal ganglia, mirroring the predilection sites of classic parenchymal NBD [2, 5, 6]. Less commonly, lobar hemispheric or cerebellar involvement has been reported [6–9]. On MRI, lesions are typically hyperintense on T2/FLAIR sequences, often associated with surrounding edema and mass effect, and show variable contrast enhancement. Diffusion-weighted imaging


***Received****: 27-02-2026 -* ***Accepted****: 24-04-2026 -* ***Published****: 01-05-2026*

**Libyan Med J.** 2026;18(5):207-210
https://doi.org/10.69667/lmj.26501
**Libyan Medical Journal**
https://lmj.ly/index.php/ojs/index *eISSN: 2079-1224*

frequently demonstrates facilitated diffusion with elevated ADC values, helping differentiate these lesions from abscesses or hypercellular tumors such as lymphoma [8–10]. Nevertheless, no single imaging feature is pathognomonic, and correlation with clinical context and treatment response is essential. A representative case from clinical practice was included to demonstrate the evolution of imaging findings and the corresponding treatment response [Figure.1- 3].

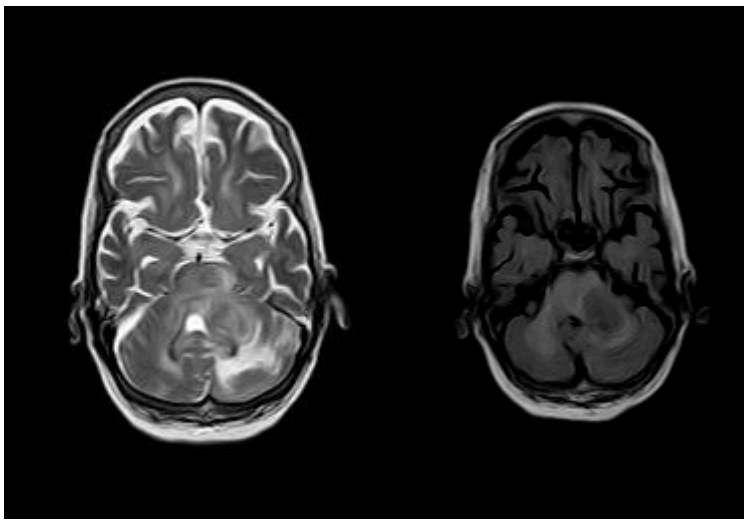

***Figure 1. Axial T2/FLAIR: High signal in the left cerebellar hemisphere and middle cerebellar peduncle***

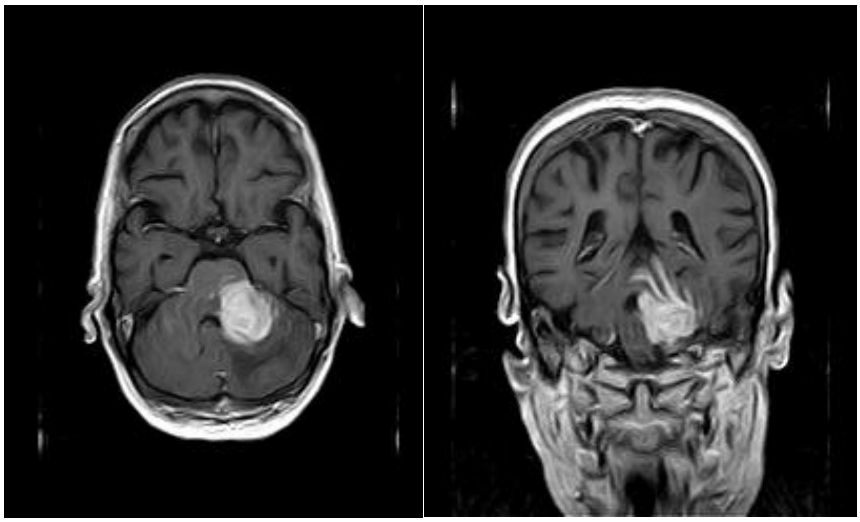

***Figure 2. Axial and Coronal post-contrast T1: Heterogeneous enhancement of cerebellar pseudomass and Extension of pseudomass into cerebellar vermis***

***Pathological Findings***

Histopathological examination, available in a subset of biopsied cases, consistently demonstrates perivascular inflammatory infiltrates, vasculitic changes, reactive gliosis, and occasional necrosis, without evidence of neoplastic cells or infection [5–7]. These findings support the concept that pseudomass lesions represent focal, intense inflammatory activity rather than true tumors.


***Received****: 27-02-2026 -* ***Accepted****: 24-04-2026 -* ***Published****: 01-05-2026*

**Libyan Med J.** 2026;18(5):207-210
https://doi.org/10.69667/lmj.26501
**Libyan Medical Journal**
https://lmj.ly/index.php/ojs/index *eISSN: 2079-1224*

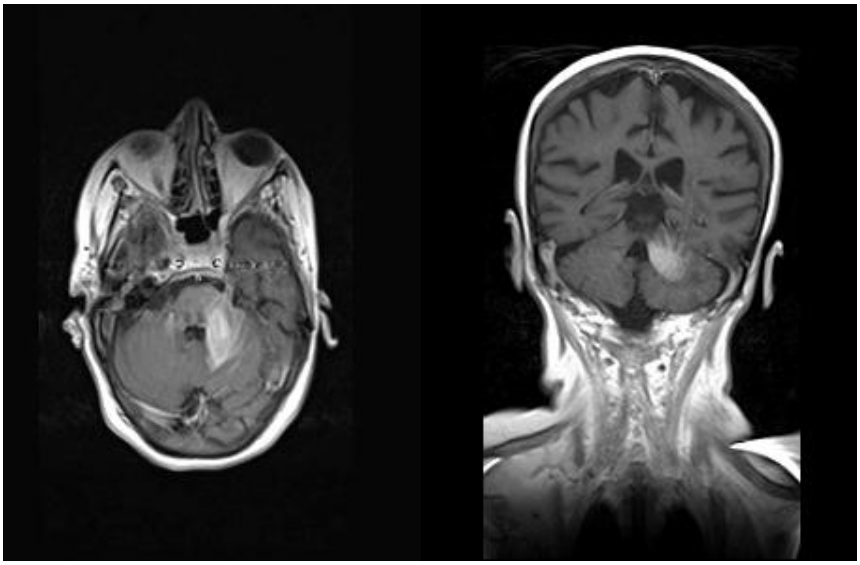

***Figure 3. Post-treatment axial T1 contrast: Partial regression of pseudomass lesion***

***Treatment and Outcomes***

High-dose corticosteroids remain the cornerstone of therapy for pseudotumoral NBD. In a 2012 review of 23 cases, nearly all patients received corticosteroids, with approximately 60% achieving complete clinical and radiological remission [5]. In a more recent review of 43 cases, immunosuppressive agents such as azathioprine or cyclophosphamide were used in approximately two-thirds of patients, with complete remission observed in about 40% [6]. Biologic therapies, particularly anti-TNF agents such as infliximab, have shown promising results in refractory or relapsing cases [6, 11]. Early recognition and initiation of immunosuppressive therapy are crucial to avoid unnecessary neurosurgical intervention and to improve neurological outcomes.

***Diagnostic Challenges and Differential Diagnosis***

The differential diagnosis of pseudotumoral NBD is broad and includes primary CNS tumors, primary CNS lymphoma, abscesses, tumefactive demyelinating lesions, and neurosarcoidosis [7–10]. Features favoring pseudotumoral NBD include subacute onset, facilitated diffusion on DWI, presence (or later recognition) of systemic BD features, and a rapid response to corticosteroids. Despite these clues, diagnostic uncertainty often necessitates biopsy in selected cases, particularly when clinical deterioration occurs, or response to immunotherapy is inadequate [5–7]. However, among the inflammatory mimickers to this observation, Tumefactive demyelinating lesions (TDLs) are the most worrisome differential diagnosis. TDLs show a typical open-ring enhancement sign and partially preserved cortical structure, and are commonly accompanied by cerebrospinal fluid oligoclonal bands, which suggests that the TDL pathology is demyelinating rather than vasculitic [12].

By contrast, pseudomass-like lesions in NBD are more frequently seen to affect the brainstem, thalamus, or basal ganglia and usually exhibit an irregular or heterogeneous enhancement pattern accompanied by atypical inflammatory CSF findings for multiple sclerosis [1]. PCNSL is an important neoplastic differential diagnosis for NBD. On MRI, PCNSL typically presents with homogeneous enhancement on contrast-enhanced T1-weighted images, a periventricular location preference, and high cellularity corresponding to inhomogeneous diffusion restriction. In contrast, histopathology in PCNSL shows compact infiltration with malignant lymphoid cells, while NBD is associated with perivascular inflammatory necrosis [13]. An accurate differentiation requires a multimodal approach combining neuroimaging features, cerebrospinal fluid analysis, clinical course, and systemic involvement to prevent misdiagnosis and unnecessary surgical intervention.

**Conclusions**

Pseudomass-like CNS lesions represent a rare but increasingly recognized manifestation of neuro-Behçet's syndrome. Their ability to mimic neoplastic and infectious processes poses significant diagnostic challenges. Awareness of this entity, careful integration of clinical and radiological findings, and early initiation of immunosuppressive therapy are essential to optimize outcomes and prevent unnecessary surgical intervention. Larger multicenter studies and registries are needed to better define the natural history, optimal treatment strategies, and long-term prognosis of pseudotumoral NBD.


***Received****: 27-02-2026 -* ***Accepted****: 24-04-2026 -* ***Published****: 01-05-2026*

**Libyan Med J.** 2026;18(5):207-210
https://doi.org/10.69667/lmj.26501
**Libyan Medical Journal**
*https://lmj.ly/index.php/ojs/index eISSN: 2079-1224*

## References

1. Kalra S, Silman A, Akman-Demir G, et al. Diagnosis and management of neuro-Behçet's disease: international consensus recommendations. J Neurol. 2014;261(9):1662-1676.
2. Akman-Demir G, Serdaroglu P, Tasçi B. Clinical patterns of neurological involvement in Behçet's disease: evaluation of 200 patients. Brain. 1999;122(11):2171-2182.
3. Al-Araji A, Kidd DP. Neuro-Behçet's disease: epidemiology, clinical characteristics, and management. Lancet Neurol. 2009;8(2):192-204.
4. Siva A. Behçet's syndrome and the nervous system. J Neurol. 2001;248(2):95-103.
5. Noël N, Bernard R, Wechsler B, et al. Pseudotumoural presentation of neuro-Behçet's disease: case series and review of the literature. Rheumatology (Oxford). 2012;51(7):1216-1225.
6. Hadj Taieb MA, Kacem I, Ben Djebara M, et al. Pseudotumoral neuro-Behçet's disease: case series and literature review. Acta Neurol Belg. 2024;124(2):431-445.
7. Bilge NS, Saylisoy S, Kasifoglu T, et al. Mass-like lesions as a rare form of neuro-Behçet's disease. Eur J Rheumatol. 2014;1(1):34-38.
8. Heo JH, Lee ST, Chu K, et al. Neuro-Behçet's disease mimicking multiple brain tumors. J Neurol Sci. 2008;264(1-2):177-181.
9. Appenzeller S, de Castro R, Queiroz L, et al. Brain tumor-like lesion in Behçet disease. Rheumatol Int. 2006;26(6):577-580.
10. Tuzgen S, Kaya A, Erdincler P, et al. Neuro-Behçet's disease mimicking cerebral tumor. Neurol India. 2003;51(3):376-378.
11. Desbois AC, Addimanda O, Bertrand A, Deroux A, Pérard L, Depaz R, Hachulla E, Lambert M, Launay D, Subran B, Ackerman F, Mariette X, Cohen F, Marie I, Salvarini C, Cacoub P, Saadoun D. Efficacy of Anti-TNFα in Severe and Refractory Neuro-Behcet Disease: An Observational Study. Medicine (Baltimore). 2016 Jun;95(23):e3550.
12. Masdeu JC, Quinto C, Olivera C, Tenner M, Leslie D, Visintainer P. Open-ring imaging sign: highly specific for atypical brain demyelination. Neurology. 2000 Apr 11;54(7):1427-33.
13. Batchelor T, Loeffler JS. Primary CNS lymphoma. J Clin Oncol. 2006 Mar 10;24(8):1281-8.


***Received****: 27-02-2026 -* ***Accepted****: 24-04-2026 -* ***Published****: 01-05-2026*